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Acute Soft Head Syndrome in Sickle Cell Disease in Qatif Central Hospital, Saudi Arabia—Case Report and Review of the Literature

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Abstract

Acute soft head syndrome or sickle cell cephalohematoma is not a common sequel or complication of Sickle cell disease (SCD). Here we describe a case report of a 15-year old Saudi male with sickle cell disease who presented with generalized vaso-occlusive crisis. 48 hours post admission, he developed generalized scalp swelling which is consistent with one of the rare complications of sickle cell disease, acute soft head syndrome. The patient was treated conservatively, without aspiration of the scalp swelling. This case draws attention that acute soft head syndrome should be one of the differential diagnoses of scalp pain and swelling in patients with sickle cell disease.

Keywords

Sickle Cell Disease, Sickle Cell Anemia, Acute Soft Head Syndrome, Sickle Cell Cephalohematoma

1. Introduction

Sickle cell disease (SCD) is one of the commonest inherited blood disorders in Saudi Arabia [1] [2]. The prevalence of SCD in Saudi Arabia varies in different regions of Saudi Arabia [2]. The Saudi Premarital Screening Program estimated the prevalence of the sickle cell gene in the adult population about 4.2% for sickle-cell trait, and for SCD was about 0.26%. The highest prevalence was noted in the Eastern province, 17% was for sickle-cell trait and 1.2% was for SCD [3]. SCD is an autosomal recessive disorder of hemoglobin that results in production of abnormal hemoglobin S due to amino acid substitution of valine for glutamic

acid. This presence of hemoglobin S leads to unstable red blood cells, with shortened survival and becomes sickle-shaped. In fact, SCD is associated with a high rate of mortality and morbidity [4].

SCD may present acutely as vaso-occlusive phenomena (including painful crisis of the bones and joints), infections, or severe anemia. It may lead to chronic complication which could involve many organ systems. The clinical feature of SCD in our region carries a milder form of the disease [5]. Avascular necrosis, and splenic complications, and Acute Chest Syndrome are exceptions [6]. Furthermore, it has been reported that sickle cell patients in the Eastern province, Saudi Arabia have persistent splenomegaly (50% - 80%) with a higher incidence of splenic sequestration and infarction [2]. 20% of them required splenectomy. The risk of avascular necrosis is 27% [2]. Sickle cell disease patients from eastern province have higher total and fetal hemoglobin, deletional α-thalassemia, and lower hemoglobin.

A2, mean cell volume, platelet counts, and reticulocytes [2]. These characteristics could explain the relatively benign phenotype of SCD in this region.

Bone and joint complication are the most common manifestation of SCD [7] [8]. The bones in sickle disease patients can be affected in several ways. It could be as bone infarction, or bone marrow hyperplasia, or osteomyelitis [7] [8]. These manifestations could involve the long bones as well as flat bones, including the skull [7]. Acute soft head syndrome is not a common sequel or complication of SCD [9]. It is also called sickle cell cephalohematoma, or skull hematoma.

Here, we describe a case that was admitted to Qatif Central Hospital and developed severe acute soft head syndrome.

To the Editor: we present a case report of A 15 year-old-Saudi male, originally from Eastern province in Saudi Arabia. He is a known case of Sickle Cell Disease (SCD), Glucose-6-Phosphate Dehydrogenase Deficiency (G6PD), α2-thalassemia, and hypersplenism. He was admitted through Emergency Department with chief complaint of lower limbs pain of 3 days duration. No history of joint swelling, fever, headache, or dizziness. There was no history of prior head trauma, or injury. Systemic review was unremarkable. His past history was positive for recurrent vaso-occlusive crisis, and one attack of scalp swelling. He was admitted to Intensive Care unit during his early childhood for central nervous system crisis, in form of stoke in which he was put in prophylactic transfusion program for few years, and then shifted to hydroxyurea, and stopped by himself.

His initial examination revealed a conscious, alert, and oriented patient. He was pale, but not jaundiced. His vital signs were: Temperature: 37.0˚C, pulse rate: 102 b/m, O_2 saturation: 98% (in room air), Blood Pressure: 120/80mmHg. No noted scalp swelling. His abdominal examination showed splenomegaly (3 cm below costal margin). Other systemic examination was unremarkable.

His initial investigations (**Table 1**) showed anemia, and mild thrombocytopenia, which were similar to his previous baseline Complete Blood Count (CBC) results. Also, he has high LDH and AST. He was managed with intravenous fluid

and narcotic analgesics.

48 hours post admission; he developed severe headache, high-grade fever, and palpitation. His examination showed a young pale male. His vital signs: temperature of (39.3˚C), pulse rate 125 b/m. His scalp examination revealed diffuse, boggy, tender, and soft swelling of the whole scalp. There was no periorbital involvement. There were no meningeal signs. The rest of his examination was unremarkable except for splenomegaly. His repeated investigations (Table 1) showed an acute drop of Hemoglobin to 7.7 gm/dL which could be explained by acute hemolysis. Complete septic workup, including blood cultures were sent, and he was covered empirically with Ceftazidime I.V. He received 2 units of packed red blood cells. His post blood transfusion Hb was 8.2 gm/dl. His blood culture came to be negative later on.

Skull X-ray was done and showed posterior calcification.

Computerized tomography (CT) of the scalp showed bilateral thick tentorial calcification; diffuse widespread swelling of the scalp that involves occipital, frontal, and temporoparietal bilaterally (Figure 1). Magnetic resonance imaging (MRI) showed acute cephalo-hematoma, no masses, midline shift, or ventricular system changes (Figure 2).

Based on his clinical presentation and imaging studies, he was diagnosed as Acute Soft Head Syndrome. He was treated conservatively with I.V fluids and analgesics. He was evaluated by a neurologist, who agreed with our management.

His scalp swelling decreased in size gradually and resolved completely. He was discharged after 9 days of admission in satisfactory condition.

Table 1. Laboratory data.

Variable	Reference range	On admission 1 day	3rd day of admission	On discharge
WBC	(4 - 10) × 10^3/uL	8.5	7.6	5.3
HBG	(13 - 17) gm/dL	10	7.7	8.1
HCT	(49 - 54)%	28.6	21.7	19
MCV	(78 - 96) fL	54.8	52.7	53.1
MCH	(27 - 32) PG	19.2	18.7	17.9
MCHC	(31.5 - 34.5) g/dL	35	35.5	33.7
RDW	(11.6 - 14)%	20.3	18.8	20
Platelet count	(150 - 340) × 10^3/uL	107	99	98
Bilirubin (total)	(0 - 1.2) mg/dL	1.29	1.3	0.8
Bilirubin (conjugated)	(0 - 0.3) mg/dL	0.26	0.4	0.2
ALT	(30 - 65) U/L	34	27	47
AST	(15 - 37) U/L	76	39	32
LDH	(81 - 230) U/L	901	751	550

Renal function test and coagulation studies all within normal range. Blood culture was negative, no growth of organisms had been found.

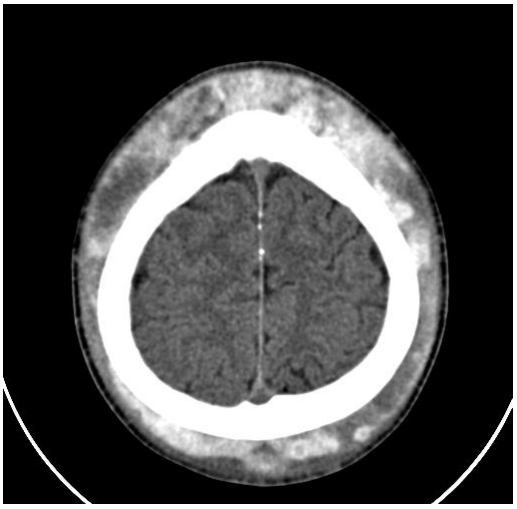

Figure 1. Scalp CT showing diffuse widespread swelling of scalp and bilateral thick tentorial calcification.

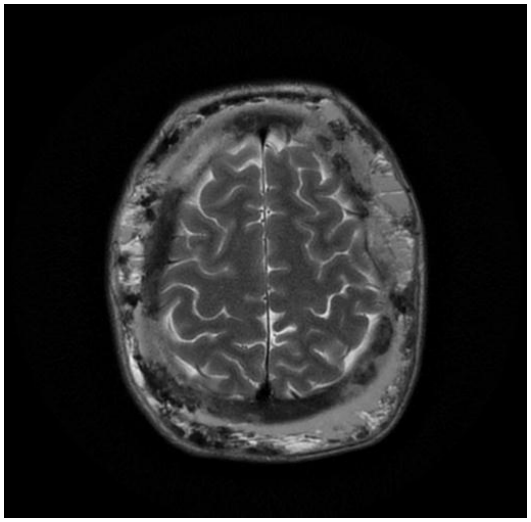

Figure 2. MRI/T2 showing stable acute cephalo-hematoma.

2. Discussion

Acute soft head syndrome is a rare manifestation of SCD [8]. This phenomenon can be presented as diffused or localized swelling [7]. Our patient presented with diffuse swelling of the whole scalp and forehead without periorbital involvement. The pathogenesis of acute soft head syndrome is most likely related to vaso-occlusive crisis, leading to skull bone marrow infarction, and tearing of small vessels [8] [9]. The process of cortical bones thinning is due to expansion of intramedullary hematopoietic tissue results in disruption of inner and outer skull margins [8]. Vascular damage is due to vasoconstriction, which will lead to obstruction of vascular lumen with sickled red blood cells, and formation of platelet fibrin thrombi due to decrease in fibrinolysis and increase activity of factor VIII. When the vascular damage is more pronounced, this will result in rupture of the vessels during crisis [7].

The differential diagnosis of sickle cell cephalohematoma, or skull hematoma must include bleeding diathesis. In our patient, all coagulation studies were within normal range.

Our patient had severe vaso-occlusive crisis for 4 days prior to the onset of the scalp swelling, a finding, which support our diagnosis. Most cases of acute soft head syndrome with SCD are managed conservatively with good hydration and analgesia. Our patient did well on conservative management. Furthermore, he was treated empirically with broad-spectrum antibiotic since it is clinically difficult to differentiate infarction from infection [7] [8]. Aspiration is not usually indicated in acute soft head syndrome unless infection is suspected [7]. Unnecessary aspiration of the scalp hematoma may lead to secondary infection. The patient was followed in the clinic after 1 month of discharge and the sickle cephalo-hematoma was resolved completely.

3. Conclusion

Acute soft head syndrome is a rare complication of SCD. Recognition of this syndrome is important for proper management. It can be managed conservatively without the need for aspiration.

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Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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